

Fate of patients with fixed subaortic stenosis after surgical removal

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SUMMARY Thirty-nine consecutive patients, aged 5 to 57 years, were followed for two to 15 years with serial haemodynamic studies after removal of fixed subaortic stenosis, which was never a "membrane". Two late deaths occurred, one sudden and one in congestive failure. Of 37 survivors, 25 were asymptomatic and could be classified as good or excellent if judged by well-being. Seven were symptomatic, two having had reoperation for fixed subaortic stenosis, and four needed long-term pacing.

Evaluation, including the effect of isoprenaline, showed important dynamic obstruction in 17, five of whom redeveloped fixed obstruction. Seven had congestive features without outflow gradients, and 14 had neither congestion nor outflow obstruction. Complete assessment therefore confirmed that only 14 (36%) were haemodynamically satisfactory; two of them had permanent pacing, and four had had aortic valve surgery.

Fixed subaortic stenosis should be removed early, when diagnosed, and completely before secondary myocardial changes occur. Patients however "well" need regular supervision and early haemodynamic assessment. The aortic valve, whether repaired, replaced, or untouched, remains a site for infective endocarditis for life.

The fixed subaortic stenosis removed at operation may not be present in that form at birth, but acquired secondary to other congenital abnormalities which remain in the patient.

Fixed subaortic stenosis, which is usually classified as a congenital lesion, takes the form of a crescentic shelf or complete ring. It is often eccentric, attached to the anterior cusp of the mitral valve and/or the left coronary cusp of the aortic valve, 1 to 2 cm below the aortic valve. Surgical excision is relatively simple and should, theoretically, relieve the haemodynamic problem. However, after surgical treatment was established, it was soon noted that results were unpredictable.¹⁻⁴

It appeared from several different series that despite removal of the fixed part of the obstruction the cardiac muscle could continue to behave physiologically like hypertrophic cardiomyopathy⁵⁻⁷ and that there might be an overlap between the conditions.

The clinical and haemodynamic state of long-term survivors after resection of fixed subaortic stenosis has now been examined with sequential studies. From these results it may be necessary to

change our views on pathogenesis, timing, and technique of surgery.

Subjects

In the National Heart Hospital between 1964 and 1977, 40 patients, aged between 5 and 57 years at the time of operation with cardiopulmonary bypass, had excision of a fixed (discrete) subaortic stenosis.

Normothermic cardiopulmonary bypass through a median sternotomy was used with partial or total coronary perfusion. The ascending aorta was opened through a vertical incision and the aortic valve was inspected and then retracted to expose the subaortic area. Valvotomy (3) or excision (4) before replacement was done when necessary. The ring or shelf was then excised completely, taking care to avoid damaging the anterior cusp of the mitral valve. After restarting coronary perfusion, the subvalvar muscle of the ventricular septum covered with thickened cream-coloured endocardium was inspected and usually seen to be bulging abnormally into the left ventricular outflow tract. In 34, a deep incision was made into the apex of

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the bulging septum, and in 25, a wedge of muscle was removed. In the past two years the technique has been changed in view of the late results. Now, all thickened endocardium is stripped off by blunt dissection from the bulging ventricular septum. This peels off like a skin to which the fixed obstructing subaortic shelf or ring is attached and ensures removal of all visible fibro-collagenous material. After the aortotomy had been sutured all the patients came off bypass easily except the one who died. Simultaneous needle pressures from the ascending aorta and left ventricular body were measured when the circulation was stable. The present policy is to return to bypass if the gradient exceeds 40 mmHg and resect more muscle or excise the whole root, depending on the pathological anatomy.

After operation, patients were examined at six monthly or annual intervals with repeated electrocardiograms, phonocardiograms, and chest radiographs. Most patients in the last three years had M-mode echocardiograms which will be discussed in another report.

Recatheterisation was performed one to 12 years after operation when agreed to by patient and referring physician; 11 patients had two postoperative studies and one had three. After reoperation further postoperative catheterisation was done under the same conditions. Gradients between the aorta and left ventricular body were measured at rest, on withdrawal of the catheter across the outflow tract, and after isoprenaline infusion sufficient to raise the heart rate to 130 to 160 per

minute. Left ventricular angiograms with 0.5 to 1 ml/kg Conray 420 injected slowly were recorded on biplane Elema still films or cine angiograms in the left anterior oblique or lateral and anteroposterior views. Routine aortograms were also performed to assess aortic regurgitation.

Ten patients were not reinvestigated, either because the referring physician refused (six), death (one), or the patient refused (three). The present clinical state is known in all patients.

For simplicity, the electrocardiographic changes of left ventricular hypertrophy from the resting record immediately before operation were graded 1 to 4:

Grade 1: Increased voltage of R in V6, above 30 mm, with normal ST segments and upright T waves.

Grade 2: Increased voltage and flattening of T waves/or ST depression in the left ventricular and/or anterior chest leads.

Grade 3: T inversion in the left chest leads.

Grade 4: Deep T inversion with ST depression of 3 to 8 mm in left ventricular leads.

Preoperative state of patients

CLINICAL

The age distribution of the patients at the time of operation and the year of operation are shown (Fig. 1a and 1b). The youngest was 5 years and the eldest was 57 years; 20 were in the second decade (23 male and 17 female). Twenty-seven had symptoms and 13 were symptom free. Left ventricular

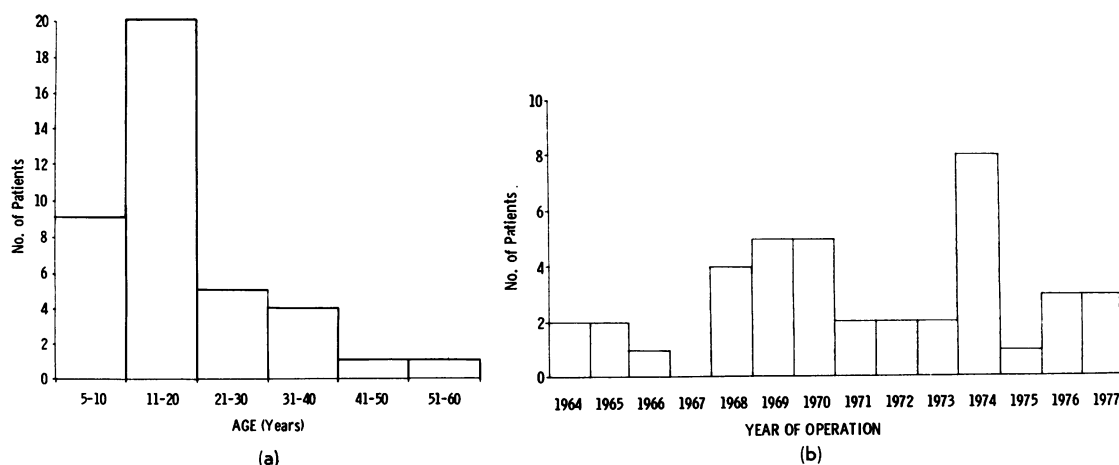
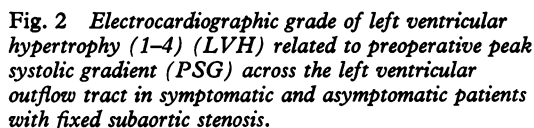


Fig. 1 (a) Age when fixed subaortic stenosis removed in 40 patients. (b) Calendar year when fixed subaortic stenosis removed in 40 patients.

The initial indications for removal of the subaortic stenosis were electrocardiographic changes of left ventricular hypertrophy (grade 1 to 4) and/or the demonstration of a peak systolic gradient at rest above 50 mmHg, symptoms in association with either of the previous criteria, or failure from associated aortic regurgitation or large duct. In the past three years, fixed subaortic stenosis was removed when diagnosed and confirmed by angiography and gradient measurement, irrespective of severity or whether or not there were associated electrocardiographic changes.



Important cardiac enlargement on the chest radiograph (CTR > 55%) was seen in five patients: the only two patients with a left-to-right shunt from a duct and failure, two patients over the age of 40 years, and the 29-year-old with a haemolysing Starr valve. A pronounced left ventricular contour at the apex was obvious in most, but low (caudal) post-stenotic dilatation of the ascending aorta was only seen in seven patients. The chest radiograph was not helpful in differentiating subaortic stenosis from other forms of congenital aortic stenosis.

At preoperative catheterisation resting gradients were measured in 34 patients. The gradients were found to be labile, being lower with general anaesthesia (six), higher with agitation, and increasing conspicuously with isoprenaline in three with gradients below 60 mmHg. Isoprenaline was not given to others before operation. The gradient increased after a single ectopic beat with an increase in both aortic and left ventricular systolic pressures as in fixed obstruction. The end-diastolic pressure in the left ventricle was raised (15 to 20 mmHg) in four patients, two with signs of heart failure and the other two with breathlessness. Unfortunately, no haemodynamic studies were done in the two older patients with established left bundle-branch block; each had been referred for aortic valve replacement despite the absence of radiological calcification.

In 19, the subaortic obstruction was difficult to see, and in 10 a "membrane" was seen on *x*-ray films. Left ventricular septal muscle and free wall muscle was grossly and irregularly thickened in 16 patients (Fig. 3a and b). Regular concentric hypertrophy with septal hypertrophy was obvious in 28 (Fig. 4A and B). The aortogram showed characteristic slow opening of the aortic valve cusps which were visible, in the absence of congenital fusion, in mid-to late systole, with uneven concavity towards the lateral. Clinically unsuspected fixed subaortic stenosis was diagnosed correctly from the aortogram in three patients from the characteristic appearances of the aortic valve movement (Fig. 5a). Doming of a mildly stenosed aortic valve above a subvalvar obstruction was recognised in seven; thickening of the valve was frequently noted. Aortic regurgitation was confirmed in 38 patients and frequently outlined the fixed subaortic obstruction (Fig. 5b) better than left ventricular angiography. The ascending aorta was dilated in 15, small in 23, and associated with slight supravulvar waisting in 12.

PREGNANCY AND FAMILY HISTORY

Three patients who also had a duct were products of pregnancies complicated by rubella. One other patient's mother was diabetic. There was a family

history of congenital heart disease in first degree relatives in two; in one a brother died suddenly with hypertrophic cardiomyopathy and another has a daughter with a long aortic ejection systolic murmur. A specific family study was not made.

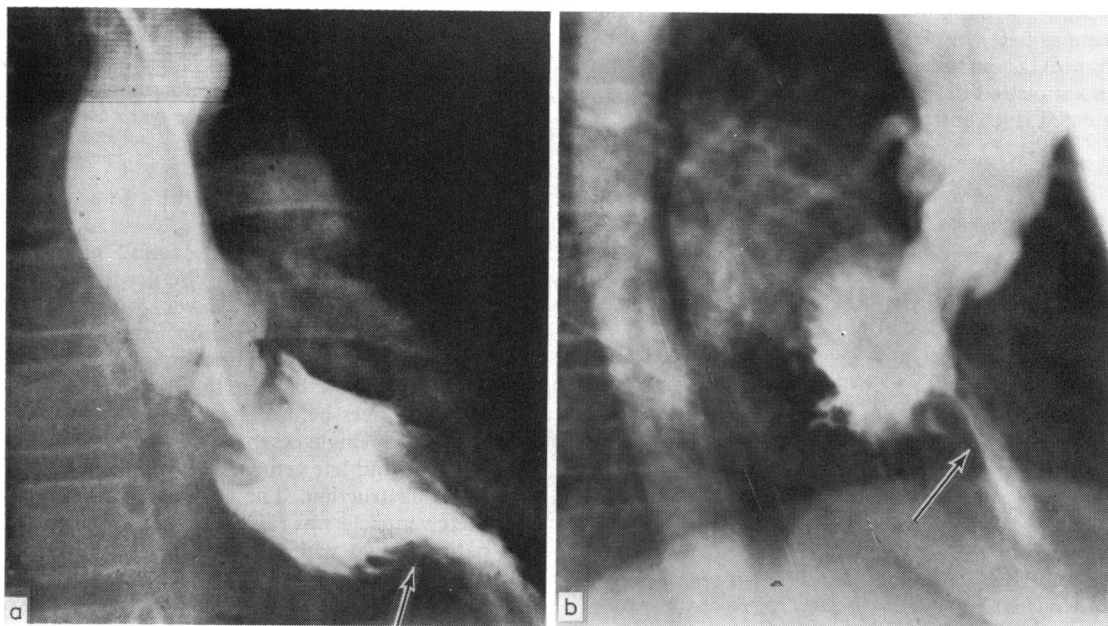


Fig. 3 Preoperative left ventricular angiograms from boy, operated on in 1974 aged 11 years, with fixed subaortic obstruction caused by crescent of tissue and thickened endocardium over severely bulging ventricular septum. Large lumps of eccentrically hypertrophied muscle are present (arrows). This patient maintained clinical and haemodynamic obstruction for five years. (a) Anteroposterior view. (b) Lateral view.

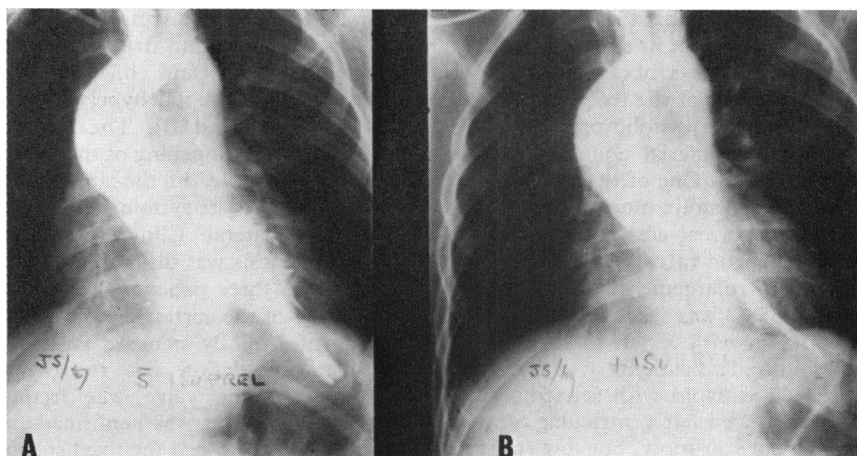
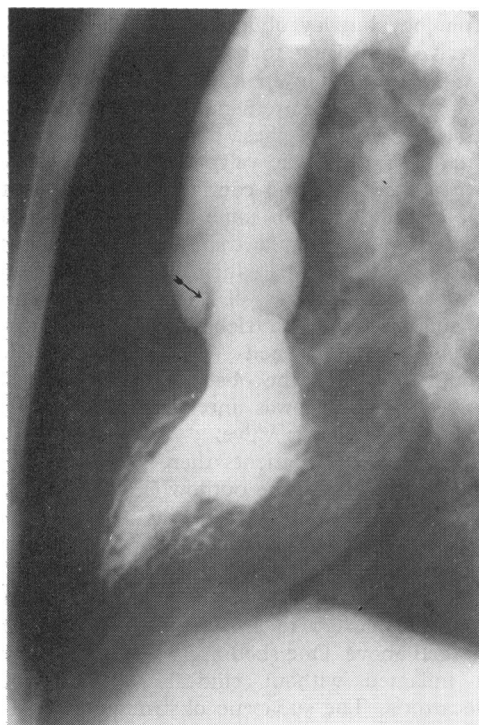
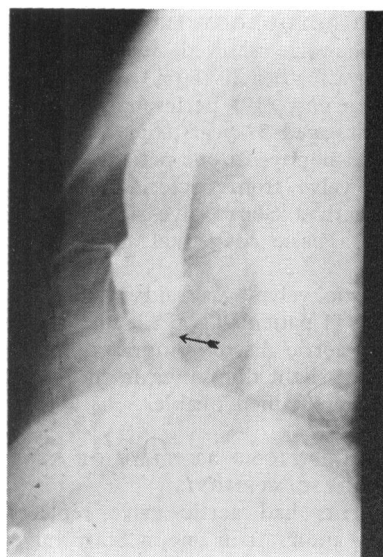


Fig. 4 Diffuse concentric hypertrophy shown on anteroposterior view of left ventricular angiogram from 6-year-old boy with fixed subaortic stenosis, grade 4 LVH and peak systolic gradient 70 mmHg. (A) Before isoprenaline. (B) After isoprenaline infusion. He maintained a good haemodynamic result seven years later with regression of LVH and resting gradient 0 mmHg rising to 20 mmHg with isoprenaline.



(a)



(b)

Fig. 5 (a) Systolic frame from lateral view of left ventricular angiogram showing characteristic movement and visibility of "normal" aortic valve with fixed subaortic stenosis beneath (not obvious). (b) Aortogram (lateral) from 12-year-old boy with fixed ring obstruction outlined (arrow) beneath regurgitant aortic valve.

CONGENITAL CARDIOVASCULAR LESIONS

Twenty-four patients (60%) had other congenital abnormalities in the cardiovascular system (Fig. 6). Ventricular septal defects were documented by angiography in seven patients; three closed spontaneously before the operation for the subaortic stenosis. The fibrous tissue closing the defect was attached to the subaortic stenosis which was proximal or contiguous with the ventricular septal defect, and there is evidence to suggest that the subaortic obstruction formed or progressed in the period when the ventricular septal defect was closing. In one child the ventricular septal defect had been closed surgically six years earlier leaving the undiagnosed fixed subaortic obstruction—the child nearly died in the postoperative period and suffered myocardial damage which was probably contributory to his sudden death a year after resection of the subaortic stenosis.

Coarctation at the classic site was resected in four patients, one aged 5 months, one aged 8 months, and two at 4 years; a further two had a mild untreated coarctation. Ten patients had a documented persistent duct, ligated earlier in nine and closing spontaneously in the first decade in one.

The aortic valve was congenitally abnormal in

nine: four had a true bicuspid aortic valve, four a "tricuspid" valve with a rudimentary fused anterior commissure, and one had a small valve in which two cusps were attached higher than usual.

In only 16 patients (40%) was there no other congenital abnormality discovered in the heart or great arteries. The subaortic stenosis had the same morphology in those with and those without congenital abnormalities.

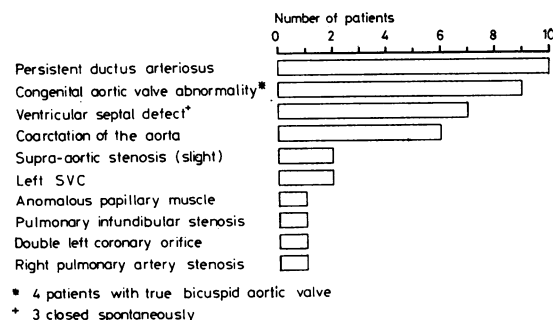


Fig. 6 Associated congenital abnormalities in the cardiovascular system in 40 patients with fixed subaortic stenosis.

ACQUIRED PATHOLOGY IN AORTIC VALVE

The aortic valve showed acquired pathological changes in a further 20 (50%); thickening of cusp edges or one cusp (19), perforated cusps (3), and in one patient, aged 57 years, the valve was seen to be thin and floppy. In one patient aged 29 years a destroyed valve from bacterial endocarditis was replaced with a Starr valve—it is not known if there was a basic congenital abnormality in this patient.

The aortic valve was said to look "probably normal" in 11 patients (27.5%), aged 5 to 32 years, despite an aortic diastolic murmur in nine. Most of these had slight thickening in the centre of one cusp which remained pliable.

OTHER OPERATIONS AT TIME OF RESECTION OF SUBAORTIC STENOSIS

Four patients had aortic valve replacement: a pulmonary autograft in one, a Starr valve in one, and a "fresh" aortic homograft in two. Other operations included closure of ventricular septal defect in three, open aortic valvotomy in three, resection of right-sided fixed ring infundibular stenosis in one, ligation of persistent ductus arteriosus in one, and relief of supra-aortic stenosis in one. The 12-year-old girl in failure with a duct and slight coarctation and a tight fixed subaortic obstruction developed ventricular fibrillation during the thoracotomy for the duct and so the incision was extended and bypass instituted to resect the subaortic obstruction after duct closure.

ANATOMY OF SUBAORTIC STENOSIS

Conventional classifications used by others^{3,4} have not been used here as we believe them to be a source of confusion and misinterpretation. Firstly, because there is little evidence that the lesion is "discrete" except in the minds of radiologists and the surgeon who resects it, and a membrane by definition "a thin pliable sheet-like tissue"* was never encountered. A simple description of what was found has been used. Whether it was near or separated from the aortic valve depended in part on its completeness, its obliquity, and the distribution and hypertrophy of muscle bordering the outflow tract. Since only one died, there was no opportunity to measure and quantify this aspect which has recently received attention,⁸ and is likely to be relevant to pathogenesis and postoperative evolution of the lesion.

Eighteen had a complete ring, which could be described as a collar. This was excised *in toto* in seven (Fig. 7a). In three, the whole ring was close to the aortic valve but in the rest it had the same

* Shorter Oxford English dictionary, 3rd edition.

morphology but lay obliquely and not parallel to the valve, appearing to be 0.5 to 1.5 cm beneath the valve, depending upon which part of the aortic valve ring the measurements were made from or assessed. In most of these oblique rings only part was close to the aortic valve lying in the "space" between the aortic and contiguous anterior mitral cusp. In others and in some with crescents, there appeared a wider area than usual between the aortic and mitral cusps which may have accounted for the apparent lowness of the obstruction. Excessive bulging of the ventricular septum, frequently covered with thickened "wash leather" endocardium, to which the obstruction was attached, occurred in 15, and was unrelated to proximity of the ring to the aortic valve.

In the other 22 patients there was a crescentic shelf projecting into the outflow (Fig. 7b); 19 had severe septal bulging. There was calcium in the crescentic web in one girl aged 14 years and in only three was the crescent close to the aortic valve. In the rest it was oblique and separated from the valve ring by about 0.5 to 1.5 cm, as were the rings described above. One shelf appeared to be vascular and inflamed without clinical signs of active endocarditis. The subaortic obstruction was often rolled and uneven with areas of increased denseness in both the crescents and rings.

HISTOLOGY

The obstructing ring or shelf was usually referred to as "fibrous". It was, in fact, a composite of different tissue cells varying from patient to patient. Often there was a large percentage of irregularly orientated acellular dense collagen fibres. Elastic fibres, thin, short, and irregularly arranged, were

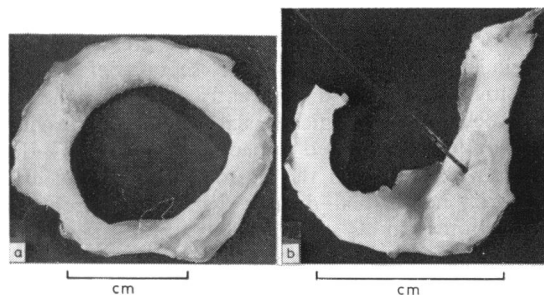


Fig. 7 (a) Complete ring removed from beneath the aortic valve from a 21-year-old man. The angiogram was reported to show a "typical membrane". (b) Crescentic shelf forming subaortic obstruction, attached to thickened endocardium stripped off the bulging ventricular septum, from a 13-year-old girl.

seen and these varied in density and distribution in different blocks from the same patient. Three patients had a large amount of elastic tissue.

In the majority of sections, elongated nuclei resembling fibroblasts were scattered sparsely, and in some sections they merged into large acellular areas and the nuclei became shorter and plump within cytoplasm which, with trichrome stains, had the staining characteristics of smooth muscle. Whether there was a ring or crescent, the histology was the same.

Vascularity was usually absent but occasional sections showed a rich network of capillaries with some arterioles containing an elastic network.

Biopsies of septal muscle were small and showed mild to moderate interfascicular fibrosis with simple hypertrophy. Classic dysplasia of the myocardium in wide areas was not seen.

Results

DEATHS

One operative death occurred in a 29-year-old man with a previous Starr valve replacement for an infected aortic valve elsewhere. At emergency reoperation a tight fixed subaortic fibromuscular stenosis was removed from beneath the Starr valve but the heart failed to come off bypass.

Two late deaths occurred in the 39 survivors; one man died aged 49 years when a ring subaortic stenosis was removed, though he had left hospital symptomatically improved but with persistent left bundle-branch block and cardiomegaly; he died five years later in congestive heart failure after the onset of atrial fibrillation. Neither a necropsy nor postoperative haemodynamic studies were done. The other death was in an asymptomatic boy aged 14 years, one year after operation, during a game of football which he had been advised not to play. Postoperative studies one year after operation showed that the left ventricular end-diastolic pressure was 19 mmHg, with neither outflow gradient nor aortic regurgitation. Unfortunately, the coroner did not consider there was need for necropsy.

SURVIVORS

The state of 37 survivors two to 15 years after operation has been summarised (Fig. 8). Twenty-five had no symptoms, considered themselves well, and were undertaking full normal activities. Five had symptoms, either a return or persistence of angina or dyspnoea, and two had paroxysmal tachycardia. Four had permanent pacemakers which required the usual supervision and changes.

Three types of physiological behaviour were

found in each group of patients and designated as dynamic obstruction, "congestive", or normal. Patients were considered to have dynamic obstruction when isoprenaline caused a gradient across the left outflow tract greater than 60 mmHg, together with characteristic features suggestive of obstruction such as long ejection systolic murmur, jerky pulse, and clear aortic second sound. Since no patient had significant aortic valve or supravalue stenosis, the dynamic obstruction was assumed or shown to be at subvalvar level, usually over a diffuse area. Fixed obstruction was accepted when resting gradients were above 50 mmHg, always associated with a high isoprenaline response, physical signs of subaortic stenosis, and a visible obstruction on angiography (Fig. 9a and b).

Patients were classified as having a "congestive" myopathic left ventricle when the end-diastolic pressure in the left ventricle exceeded 10 mmHg (pre A) with low ejection fractions below 55 and/or radiological, and/or symptomatic features of congestion. Patients evaluated as truly good ("normal") (14) had none of the above haemodynamic findings; there was maintained improvement of the electrocardiogram, absence of symptoms, and no severe aortic regurgitation.

Fixed obstruction—reformed and reoperated

Fixed obstruction appeared two to nine years after the first operation in four patients and is probably present in a fifth. The four were operated on by the same surgeon who found again thickened endo-

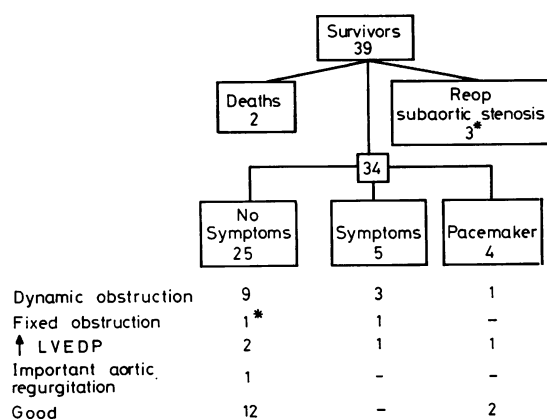


Fig. 8 Summary of state of 39 survivors two to 15 years after resection of fixed subaortic stenosis. Correlation with haemodynamic state. LVEDP, left ventricular end-diastolic pressure; *, patient operated on after paper accepted for publication

cardium attached to a protruding shelf which had previously been excised. The dynamics of these five patients have been summarised (Table 1). The electrocardiogram worsened after initial improvement. Three were symptomatic.

At the first cardiac catheterisation after operation

all had an intense response to isoprenaline. In case 1 the obstruction probably had not been adequately relieved, as a long muscular tunnel persisted allowing a resting gradient of 83 mmHg one year later. The patient had been in failure at the time of the first operation because of an associated large duct.

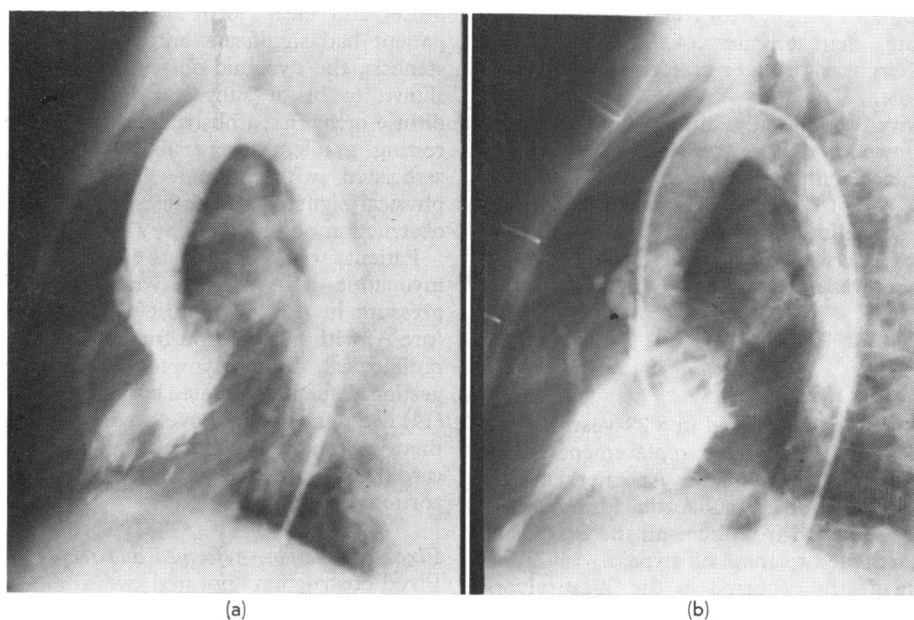


Fig. 9 Lateral views of left ventricular angiograms before a fixed subaortic ring was resected (a), and five years later when a tunnel obstruction had developed (b). After the first operation the resting gradient across the outflow was 20 mmHg, the aortic valve was normal, and the electrocardiogram improved. Four years later syncope and angina returned with worsening of the electrocardiogram. At reoperation the fixed shelf had reaccumulated over a long fixed fibromuscular obstruction.

Table 1 Peak systolic gradients across the left ventricular outflow tract before and after operation in five patients who redeveloped fixed subaortic stenosis

Case no.	Age at opn.	Other lesion	Peak systolic gradient (mmHg)					Age at reop.	Peak systolic gradient after second operation			Comment
			Preop. rest	Immediately postop.	Year	Rest	Isoprenaline		Year	Rest	Isoprenaline	
1	12	Aortic stenosis, duct, mild coarctation	80	—	1½	83	—	15	1	20	80	Re-resection of subaortic stenosis LVEDP ↑
					1	25	140		3	8	8	
2	11	—	120	30	9	55	125	20	6/12	5	50	Aortic root replaced
					2	18	56					
3	10	—	60	10	4	50	100	15	—	—	—	Aortic root replaced
					1	85	185					
4	15	—	90	50	10	60	150	—	—	—	—	Aortic root replaced
					1	40*	80*					
5?	11	—	80	5	9	50	90	—	—	—	—	

* Catheterisation performed under general anaesthetic.

LVEDP, left ventricular end-diastolic pressure.

It is possible that the potential for persistent outflow obstruction had been underestimated at the hurried operation when the left ventricle was dilated after failure and ventricular fibrillation. As the heart, relieved of the volume overload, recovered tonicity the outflow became more obstructed and the fixed shelf part redeveloped quickly as the result of continued persistent turbulence. One other (case 4) had an 85 mmHg resting gradient at the first study, having had a crescent initially removed. At reoperation there was slight fusion of aortic valve cusps and intense subvalvar endocardial thickening and protrusion of a shelf into the outflow. Aortic root replacement with reimplantation of the coronary arteries was done.

The other two patients did not have high resting gradients at the first postoperative catheterisation and so it is difficult to claim that important fixed obstruction had been left behind. Certainly abnormal muscle remained in all and the ascending aorta was not dilated and had a slight supra-aortic narrowing of no haemodynamic significance in three of the five.

In three patients further resection of the subvalvar muscle and redeveloped shelf did not relieve the obstruction and high gradients between the left ventricle and ascending aorta persisted. The whole aortic root was then excised and replaced with a homograft aortic root and valve, with reimplantation of the coronary arteries.⁹ This appeared to relieve the obstruction successfully, since postoperative studies six to 12 months later confirmed resting gradients 5 to 20 mmHg, rising to 50 to 80 mmHg

with isoprenaline stimulation. Case 1 with a stimulated gradient of 80 mmHg did not have a root replacement.

The fifth patient (case 5) probably has a redeveloped fixed obstruction but it is not visible on the recent angiogram despite the high pressures. He has been treated with propranolol and no longer has new symptoms.

Dynamic obstruction

Seventeen patients (including those who developed fixed obstruction) had haemodynamic evidence of dynamic obstruction and/or physical signs suggesting its presence. Intravenous propranolol in seven altered the signs, making the jerky pulse become normal and diminishing the length of the ejection systolic murmur.

The relation to the age when operated on, the severity of the preoperative electrocardiographic hypertrophy, the postoperative peak systolic gradient measured at operation after resection and at the first postoperative study, and whether or not they had dynamic obstruction are shown (Fig. 10a and b).

The dynamic obstruction appeared to persist more often in those patients who had grade 4 left ventricular hypertrophy before operation. The measurement of the gradient immediately after resection, on the table, did not reflect the subsequent course though those left with gradients over 25 mmHg were more likely to have dynamic obstruction.

Four patients developed angina, syncope, or

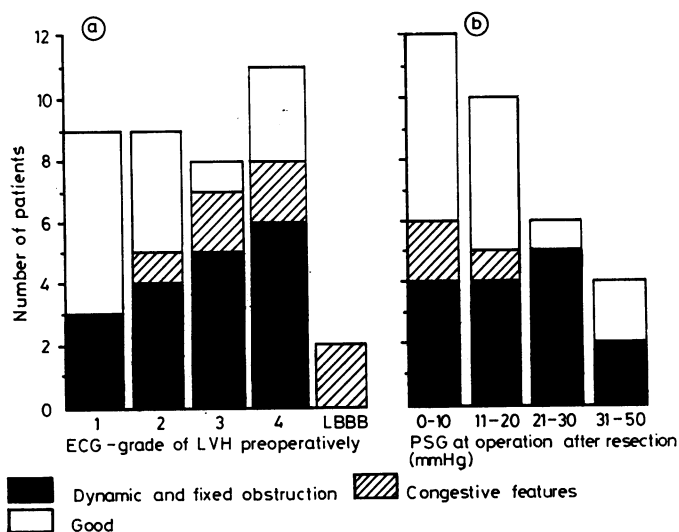


Fig. 10 Correlation between grade of left ventricular hypertrophy present before operation (a) and peak systolic gradient measured immediately after resection of subaortic stenosis on the table (b) and the subsequent findings of dynamic obstruction, congestive myopathic ventricle, or normal dynamics.

dyspnoea which reappeared after a period without symptoms. One who required permanent pacing and studies before implantation of the pacemaker showed how isoprenaline produced a high gradient (Fig. 11a), but when the heart rate was increased by pacing it lowered the gradient (Fig. 11b). Thirteen patients with dynamic obstruction were symptom free and considered themselves well, but in four the electrocardiograms were deteriorating after five years.

Repeated haemodynamic studies showed that in seven of eight patients with high gradients after isoprenaline, the response persisted two to 10 years later (Fig. 12). In one, the resting gradient and response to isoprenaline had diminished eight years later. Three patients with minimal response to isoprenaline remained the same six to 12 years later.

Redevelopment of a fixed obstruction occurred as frequently in those where a ring had been excised as in those with a crescent. Important septal thickening and dynamic bulging was seen in 32 and postoperative dynamic obstruction occurred in 15 of these. However, in seven where the septum was not considered to be unusually bulging and thick, two developed severe late dynamic obstruction with symptoms.

Now, patients with dynamic obstruction and

appropriate clinical signs are treated with maintenance propranolol 20 to 40 mg bd. It is too soon to assess the long-term results of this.

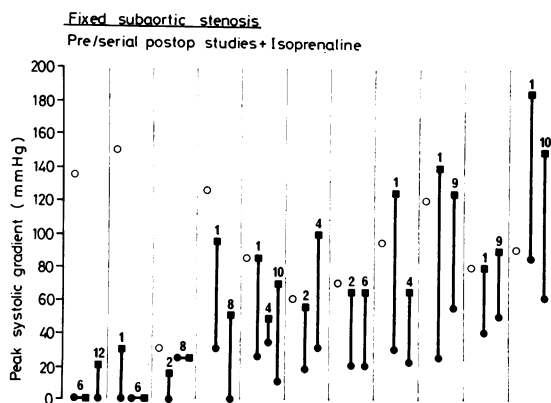


Fig. 12 Peak systolic gradients across the left ventricular outflow tract at rest before operation ○, at the first cardiac catheter at rest ●, and after isoprenaline ■, and at subsequent studies in 11 patients who had repeated catheterisation after resection of fixed subaortic stenosis. The number above each vertical line is the postoperative year at the time of study.

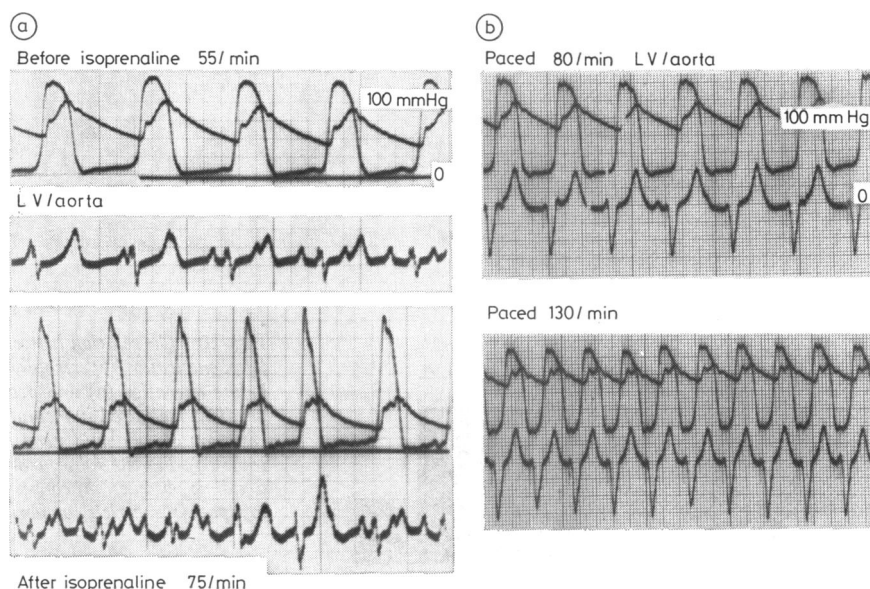


Fig. 11 Left ventricular outflow gradients measured four months after resection of fixed subaortic stenosis and closure of ventricular septal defect in patient who developed complete heart block at operation.

(a) Rest. In block (rate 55/min), gradient 50 mmHg. Below. After isoprenaline (rate 75/min), gradient 110 mmHg.

(b) Paced 80/min, gradient 25 mmHg. Paced 130/min, gradient 20 mmHg.

Congestive group

Seven patients had "congestive" features. Two died and one developed heart block and failure six years later and still has cardiomegaly and mild congestive failure. One, a woman aged 38 years, has dyspnoea which is improved by diuretics. One boy aged 11 years at operation was in severe left ventricular failure from an added duct. Ten years later his heart remained large (ejection fraction 55 per cent), with moderate aortic regurgitation but he was improved and symptom free. Two other patients aged 15 and 21 years appeared two to 12 years later with progressive mitral regurgitation and mild congestive features confirmed by investigation. One was a child from a pregnancy complicated by rubella.

This group of seven included three patients over 35 years with larger hearts than those with dynamic obstruction and the two with complete left bundle-branch block before surgery. None had important residual outflow gradients.

Symptomatic

Seven patients developed symptoms and they had either dynamic obstruction (six) with redevelopment of fixed obstruction in three or congestive features in one. Two were reoperated on for fixed obstruction and became symptom free.

Good results—physiological and without symptoms

Of the 14 patients who maintained good physiological dynamics and whose electrocardiograms improved, two had permanent pacing systems and four had had aortic valvotomy or replacement. Thus, only eight patients had not required expert help over a two- to 15-year period. After reoperation a further two can now be added to this group.

Aortic regurgitation

Ten patients had no audible aortic regurgitation after operation; one had had open aortic valvotomy and one had an aortic valve replacement with a homograft—it had been present in nine of these before surgery. Nineteen had grade 1 murmurs and seven had mild regurgitation. Three, already mentioned, had moderate aortic regurgitation, one with a pulmonary autograft, another with a dilated root having had a large duct, and another after open aortic valvotomy. No patient with significant regurgitation had either dynamic or redeveloped fixed subaortic stenosis.

Mitral regurgitation

After operation four patients developed mitral regurgitation which was mild in two and moderate in two. One had trivial regurgitation before operation which increased after; she retained a high gradient

on isoprenaline two years after the removal of the fixed obstruction and had been noted to have an anomalously placed papillary muscle at operation. Mitral regurgitation was new in three, appearing one to 10 years later; none had outflow gradients with isoprenaline stimulation, and the left ventricular end-diastolic pressures were raised in all. There was no evidence of congenital lesions of the mitral valve, and it is presumed that the mitral regurgitation was secondary to left ventricular dysfunction.

Electrocardiogram

The changes in the electrocardiograms have been summarised (Fig. 13). Left ventricular voltage reduced usually by the end of the first postoperative year.

In four, the electrocardiogram improved initially and then deteriorated four to eight years later and all these had dynamic or reacquired fixed obstruction.

In the first few months the T waves became sharply inverted over the left ventricular leads

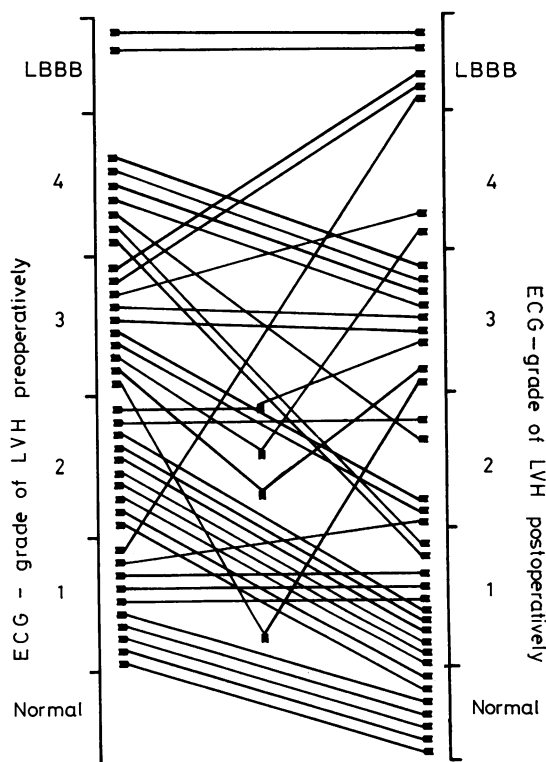


Fig. 13 Grade of left ventricular hypertrophy before and after, as assessed at most recent attendance (maximum time from surgery).

presumably in relation to myopericarditis. This usually changed after six months and so assessment of the electrocardiogram was only made after this period.

Left anterior hemiblock developed after operation in five. It was unchanged in two.

Right bundle-branch block appeared after operation in four, three of whom had the right ventricle opened.

New left bundle-branch block was caused by operation in three, aged 12 to 35 years. These all had muscle resection and had no gradients across the outflow at rest or after isoprenaline stimulation. Transient left bundle-branch block appeared for one to four weeks after operation in five.

Complete heart block

Three of 39 patients, aged 8, 16, and 31 years, acquired complete heart block at operation with heart rates of 40 to 50 per minute which were associated with cardiac failure in two.

Two of the three patients had a small ventricular septal defect closed from the right ventricle at the same time as the subaortic stenosis was removed from the outflow. One also had resection of a fixed subpulmonary stenosis.

The third patient, in whom a previously documented ventricular septal defect had closed spontaneously, developed bacterial endocarditis at the age of 31 years resulting in a destroyed aortic valve. At operation four months later a fixed subaortic stenosis was removed and the aortic valve was replaced with a homograft. The residual gradient after operation on the table was not measured; 2:1 heart block returning to sinus rhythm occurred, but three weeks after operation a lithium iodide demand pacemaker was inserted because of Adams-Stokes attacks. He returned to sinus rhythm three years later and his pacemaker is

not functioning. He has no evidence of residual fixed or labile obstruction. The heart size is normal and there is trivial aortic regurgitation.

Four years after resection, complete heart block developed in a patient aged 57 years at operation. He had left anterior hemiblock with left bundle-branch block before operation which was unchanged with prolonged PR interval after operation. He has a permanent pacemaker and is in controlled failure 15 years after operation. The state of his coronary arteries is not known.

Other rhythm disorders

Six patients had recurrent ventricular ectopics at rest, found on the routine electrocardiogram. Three of the patients had dilatation of the left ventricle with a raised end-diastolic pressure and one of these died suddenly. The other, a girl, had moderate subaortic obstruction with a gradient of 50 mmHg after isoprenaline eight years later but is otherwise well.

Supraventricular tachycardia recurred in a girl who required reoperation for fixed subaortic stenosis (case 1). She still has the attacks which are now less troublesome than previously. Another girl with Wolff-Parkinson-White syndrome continues to have short attacks controlled by propranolol 120 mg/day.

One other boy had nodal escape beats. Atrial fibrillation causing failure and death occurred in one man aged 49 years.

Pregnancies

Four patients had five successful pregnancies after operation. One had a spontaneous abortion at 10 weeks and another had a termination. None had cardiac problems during the pregnancies and all the children are normal. The postoperative haemodynamics and electrocardiographic state are shown in Table 2. The presence of labile obstruction

Table 2 *Summary of postoperative left ventricular/aortic peak systolic gradients in four patients who completed five pregnancies successfully*

Age at opn. (y)	Postop year pregnancy	Postoperative haemodynamics				Duration of follow-up (y)
		Year	Peak systolic gradient		Grade of LVH on ECG	
			Rest	Isoprenaline		
11	8	2	20	65	2	11
		6	20	65	3	
15	5, 7	1	85	185	1	11
		10	60	150		
24	2	Signs of obst + aortic regurgitation				4
12	9	1	55	140	1	9

LVH left ventricular hypertrophy; Obst, obstruction.

is not a deterrent to successful pregnancy. Propranolol was not given to any during the pregnancy.

Infective endocarditis

No patient had infective endocarditis in the postoperative period. All had careful protection for dental manipulations and extractions.

POSTOPERATIVE INVESTIGATION: SUMMARY OF FINDINGS AND CORRELATIONS

Twenty-nine patients had elective postoperative cardiac catheterisation with angiography one to 12 years after operation; 11 had repeated studies. Ten patients had no investigations for reasons already given. Peak systolic gradients before and after operation, at the first postoperative study at rest, and with isoprenaline are shown (Fig. 14a and b). The height of the preoperative gradient could not be used to predict a positive response to isoprenaline (dynamic obstruction) after operation though it was more likely in those with gradients above 80 mmHg and unlikely in those with gradients of 60 mmHg or less (Fig. 14b).

There was no absolute correlation between the height of the gradient measured immediately after operation and the subsequent haemodynamic response. Those left with gradients above 25 to 45 mmHg at operation often had a positive dynamic obstruction at the first postoperative study. However, one patient with a resting gradient of 55 mmHg immediately after resection, which remained and rose to 90 mmHg with isoprenaline, showed regression of left ventricular hypertrophy from grade 4 to 1, and nine years later the patient had no resting gradient and only 50 mmHg after isoprenaline.

By contrast, those with absent or small gradients on the table after resection usually did not have dynamic obstruction, but there were two unexpected exceptions who developed worsening electrocardiograms and severe dynamic obstruction, one with symptoms.

Only one of the seven patients with a raised end-diastolic pressure in the left ventricle before operation had the capacity to develop severe outflow obstruction in the postoperative course. This was

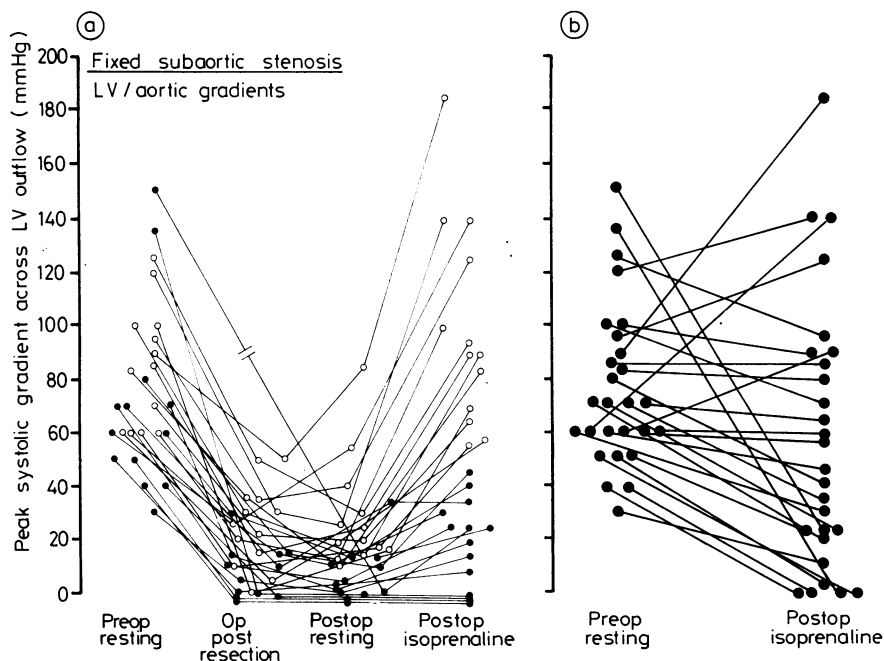


Fig. 14 Peak systolic gradients across the left ventricular outflow tract before and after resection of a fixed subaortic stenosis. (a) Shows gradients at rest before operation, immediately after resection at surgery, resting at first postoperative catheter performed one to five years after the operation, and after isoprenaline stimulation. For clarity, open circles refer to patients whose gradients rose above 50 mmHg with isoprenaline after operation. Black circles refer to patients in whom isoprenaline produced gradients of less than 45 mmHg. (b) Peak systolic gradient before operation and gradient produced by isoprenaline after operation in individual patients.

a 12-year-old girl who presented in failure with a duct.

The response of the left ventricular and aortic pressures after an ectopic beat was as seen in obstructive hypertrophic myopathy in the group with dynamic gradients but was normal in those with "myopathic" ventricles and in those with unimportant or absent outflow gradients. Angiography usually showed little change from the preoperative appearances of the left ventricular mass. The aortic valve appeared to open more normally but was usually still visible suggesting some thickening of the cusps. Angiographic assessment of postoperative fixed obstruction was even more difficult than before operation. The appearance of the left outflow tract was most normal and open in those with important aortic regurgitation and in two after total root and valve replacement with a "fresh" homograft.

Left ventricular function, as measured by ejection fractions and plotting the shortening curve, showed that the majority of patients had ejection fractions ranging between 65 and 82 per cent (H W M Plokker, personal observations). Three patients had a low ejection fraction ranging from 30 to 50 per cent, two of the three having been in heart failure before operation. None of these had a gradient above 25 mmHg nor a positive isoprenaline response.

Postoperative left ventricular angiography in 12

showed an abnormal looking left ventricle (like hypertrophic cardiomyopathy), either unchanged in comparison with preoperative appearance or worse (Fig. 15a and b). Three patients with gradients above 50 mmHg and a positive isoprenaline response, seven to 10 years after operation, had the appearance of "tunnel obstruction" (Fig. 9b) which had not been present before the initial procedure.

Discussion

Surgical removal of a fixed subaortic stenosis as a first procedure under direct vision had a low peri-operative mortality and can be considered a safe procedure unless the patient was referred in a terminal state with an added complication, as exemplified by the one operative death in 40 patients. Careful inspection of the subvalvar region in any aortic valve replacement, particularly when the valve has been attacked by endocarditis and surgical attention is centred on that, is mandatory, and had this been done at the first operation death at the subsequent operation might have been prevented.

Problems after surgery may develop in relation to damage to the aortic valve, to the mitral valve, or to the conducting tissue which appears to be vulnerable in patients who have needed additional right ventriculotomy to close a ventricular septal

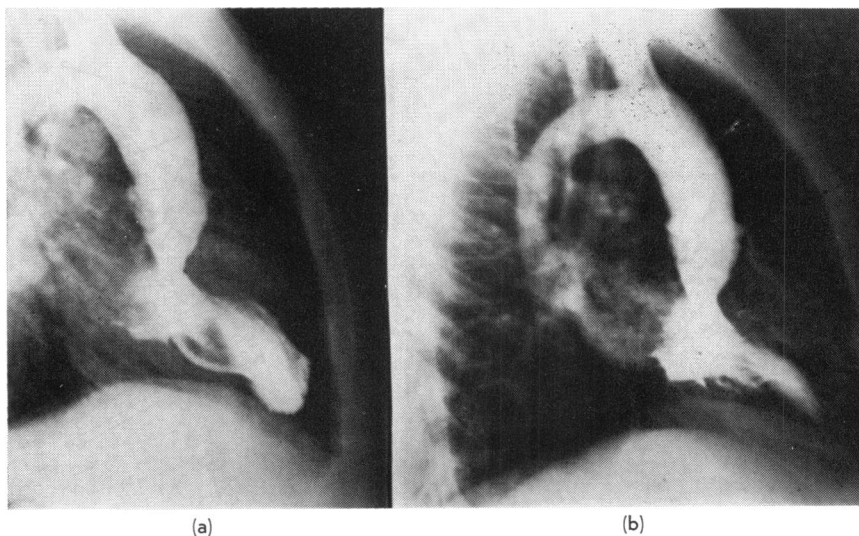


Fig. 15 Left ventricular angiography before operation (a), and one year after operation (b) showing persistence of abnormal muscle and some narrowing of the subaortic region. Immediately after resection the gradient was 5 mmHg and at first postoperative catheterisation it was 40 mmHg, rising to 80 mmHg with isoprenaline. Nine years later it was 50 mmHg, rising to 90 mmHg and he had breathlessness, with worsening of the electrocardiogram and features of persistent dynamic obstruction relieved by propranolol. Redeveloped fixed obstruction is suspected.

defect or resect infundibular stenosis. Our practice now is to warn relatives of patients with the dual problems that long-term pacing might be needed after operation. Even with stable rates of 40 to 50/min patients have symptoms, probably a result of the diffuse myocardial problem and for this as well as other sinister developments, permanent pacing is necessary once block has been established. It may also be needed if transient block has occurred.

Surgical trauma is not the only cause of complete heart block in patients with fixed subaortic stenosis. It may develop late, as in the eldest patient aged 57 years, from progressive myocardial fibrosis or associated coronary disease. Complete heart block in patients with discrete subaortic stenosis may also be acquired during endocarditis on the aortic valve from a burrowing infected aneurysm.¹¹

Pathological and physiological abnormalities of the aortic valve are present in the majority of patients with surgically significant subaortic stenosis. Lesions may be congenital (22.5%) or more commonly are acquired with thickening from jet lesions which for life remain a site for infective endocarditis before and after operation. The incidence and severity of acquired changes on the aortic valve increase with age; this is one reason for removing fixed subaortic stenosis early after diagnosis.

Aortic regurgitation sometimes increases after removal of a subaortic stenosis if a cusp is damaged or after an over-zealous valvotomy. Those in whom aortic valve replacement has been needed will require further surgery. Mild aortic regurgitation is usually unchanged after careful removal of the subaortic obstruction; the regurgitant murmur may disappear. Those with a successful aortic valve replacement, completely relieving the valvar gradient, did not develop dynamic obstruction and seemed to do better in the long term.

The anterior cusp of the mitral valve may be damaged during the resection of the attached fibrous stricture. The papillary muscle may be aberrantly attached and contribute to new or worsening mitral regurgitation. Mitral regurgitation may also develop secondary to progressive or increasing left ventricular dysfunction. Whether this is related to intraoperative ischaemia, which was not prolonged in this series, or to natural progression of myocardial disease is uncertain.

Apart from these complications which occurred in 20 per cent, it might be assumed that once the fixed organic obstruction has been removed the patient's major problem is solved. This series shows that the problem is not so simple. Lambert *et al.*¹² in assessing a small group of patients after removal of fixed subaortic stenosis commented that their state was "unpredictable" and others have

been faced with the need for reoperation and worsening electrocardiograms.^{4 13}

Persistent dynamic obstruction with and without fixed obstruction occurred in 44 per cent of survivors. Another seven (18%) had features of congestive myopathy which appeared to be progressive in one who died; in the other death the left ventricular dysfunction may have been iatrogenic.

Clinical signs initially suggested the presence of dynamic obstruction in the postoperative patient who presented with jerky pulses with rapid upstroke, persistent long aortic systolic ejection murmur, and hyperdynamic left ventricular pulsation in the absence of obvious aortic regurgitation.

One is tempted to assume that the response produced by isoprenaline may also occur with emotional and other adrenergic stresses of daily life in such prone patients and that may be the cause of electrocardiographic variation and deterioration seen in some of the late postoperative patients. Similar dynamic responses occur sometimes after surgery for other forms of left ventricular obstruction such as aortic valve stenosis, supra-aortic stenosis, and occasionally coarctation of the aorta. Large areas of dysplastic myocardium in the septum, papillary muscles, and free wall of the left ventricle have been demonstrated in these and other congenital malformations.^{14 15}

Such muscle presumably develops and behaves abnormally in the presence of fixed obstruction and remains behind after the obstruction is removed. Given the stimulus from increased work from residual obstruction, or possibly athleticism, as well as sympathomimetic drugs, such muscle may continue to function abnormally and even progress to cause symptoms. It is clear that the giving of isoprenaline support in the early postoperative period is undesirable unless the myocardium has been in failure or been insulted during the operation.

The finding of dynamic obstruction in the early years after apparently successful resection of a fixed subvalvar obstruction in more than 50 per cent of patients operated on under 20 years requires explanation and understanding. Its influence on long-term prognosis appears to be of importance from this series even though patients with it were well. Certainly, in the early years, there were no signs of symptoms or deterioration except in the one patient who clearly had residual fixed obstruction left at the first operation performed hastily after cardiac arrest and who required reoperation two years later.

The dynamic obstruction persisted in seven of the eight in whom it was initially demonstrated, and who had further studies four to 10 years later.

In only three was there a slight increase in the resting gradient whereas in the others it actually fell with time. Thus, it appears in the small group who have had repeated catheterisation that the presence of dynamic obstruction may predispose to development of progressive fixed obstruction or progressive dynamic obstruction. Neither course is predictable with certainty for there was one boy with a gradient on isoprenaline of 90 mmHg one year after operation and eight years later he had no gradient at rest, rising to only 50 mmHg with isoprenaline and an electrocardiogram originally with grade 4 changes which regressed to grade 1. Perhaps it is relevant that he led an entirely sedentary life, refusing employment. Looking at the whole group with evidence of dynamic obstruction, nine are without symptoms but the electrocardiogram has deteriorated after improvement for two to four years in three, a further three developed angina or dyspnoea, and five have a reformed fixed subaortic ring or crescent. Only six had no long-term problems for four to seven years and so it appears that despite absence of symptoms and electrocardiographic improvement, these patients may still develop problems, with or without redevelopment of fixed obstruction. In the hope of preventing progressive muscular obstruction, propranolol in small doses is now given long-term to patients with dynamic obstruction. Large doses are not given as children and adolescents seem unusually sensitive, with falling off in concentration and school performance. Pregnancy is not contraindicated in these patients; five pregnancies were successfully completed and all of the patients had important dynamic obstruction. Presumably the volume overload was beneficial for the hypercontractile ventricle.

It could be argued that part of the dynamic gradients was sometimes created by trapping of the catheter tip which may be a problem in hypertrophic obstructive myopathy. Care was taken to try to avoid this and there were clinical features suggesting the presence of dynamic obstruction in those patients where it had been documented. The possibility that these were spurious gradients has therefore been ignored.

Not all patients with fixed subaortic stenosis show signs of dynamic obstruction after operation. Since it appears to be important in the postoperative evolution, retrospective assessment of preoperative data has been looked at to see who is "at risk". The anatomy of the fixed obstruction, whether ring or crescent, was not a predictor of the postoperative course, nor did it appear to matter whether the obstruction was all close to the aortic valve or lying obliquely and deeper. Age at the

time of operation appeared to have some influence. Persistent dynamic obstruction was not seen in those below 8 years or those over 35 years. Younger patients may have had milder obstruction with less secondary effect on the myocardium; older patients perhaps had more fibrosis as was suggested by the more frequent "myopathic" response. Dynamic obstruction was most common in those operated on in the second decade which is when the majority presented. It was also more frequent, but not constant, when there was grade 4 left ventricular hypertrophy established and was never seen in those with left bundle-branch block before operation. The best indication that dynamic obstruction would persist was the morphology of the left ventricle on angiography and not the anatomy of the obstruction removed. Those with eccentric muscle lumps and excessive septal hypertrophy in systole (Fig. 3 and 15), that is some of the features described with "primary" hypertrophic cardiomyopathy including midventricular obstruction, always had postoperative dynamic obstruction. Those with concentric regular hypertrophy, even with grade 4 electrocardiographic left ventricular hypertrophy, did not develop it provided the fixed outflow obstruction was completely relieved at the initial operation.

Not only were there different angiographic appearances of the left ventricle but also the ascending aorta and root showed different forms. There were some who had no post-stenotic dilatation with small or normal sized aortic root and ring, often with slight supravalvular narrowing; these were associated with eccentric muscle, persistence of dynamic obstruction, and reaccumulated fixed obstruction. Residual resting gradients were usually higher in this group. There were others with a normal or wide root and ring; these had more smooth concentric hypertrophy or even dilated ventricles. Such observations suggest that fixed subaortic stenosis may occur in two different pathological cardiovascular systems even though the haemodynamics and clinical presentation may be the same. We suspect that the first group may have serious diffuse congenital cardiovascular disease in which areas of extensive congenital dysplastic myocardium are present^{14 15} with a small root and abnormal aorta, as occurs in variant forms of congenital aortic valve stenosis¹⁶ and some types of supra-aortic stenosis,¹⁷ and a milder more simple condition where the congenital disease is mainly localised to the subvalvar area. The latter are the ones with a better postoperative course.

Dysplastic muscle is commonly found in association with structural congenital cardiac anomalies. Whether it manifests clinically probably depends on

the extent of myocardial involvement and the basic haemodynamic disturbance. Patients with distal outflow obstruction, either on the right or left side, and excess dysplasia are almost certainly the ones who may have clinical manifestations of added problems. We suspect that a high proportion of patients with fixed subaortic stenosis are in this category. It is thus important to remove the visible fixed part of the obstruction as completely as possible in such patients. The gradients recorded on the table were not constantly helpful and often gave the surgeon false security. If gradients on the table were over 30 mmHg dynamic obstruction and further fixed obstruction might develop but if they were 0 to 20 mmHg it could also happen. This presumably relates to the state of the myocardium, temporarily poisoned or ischaemic after the bypass; such measurements may be more meaningful, with techniques of better protection of the myocardium, or after the use of isoprenaline. Patients with significant aortic regurgitation, aortic valve replacement, and no, or small, resting gradients at the first study did not develop dynamic or progressive obstruction.

When fixed subaortic stenosis reaccumulated, it presented a difficult surgical problem since re-excision of the shelf or ring alone did not relieve the gradients. Good haemodynamic relief was achieved only after excision of the whole aortic root, ring, and cusps. When this was done, together with subvalvar muscle resection, the muscular outflow opened like a flower once the small ring was removed.⁹ In three of these patients the root and valve were replaced by an adult fresh homograft into which the coronary arteries were reimplemented. Postoperative studies have confirmed complete abolition of the gradients, with a competent aortic valve. This procedure seems a more attractive and physiological answer to diffuse left ventricular outflow obstruction than the placing of an apical-abdominal aortic tube. The need for such extensive surgery for what was initially a comparatively simple lesion highlights the malignant aspects of fixed subaortic stenosis if the lesion is not treated early and completely as its continued presence in certain patients with congenital cardiovascular disease may predispose to the formation of "tunnel" obstruction.

Redevelopment of fixed subaortic obstruction is obviously related to several factors. The ability to reform the fixed subaortic stenosis which has the same histopathology as the first ring or shelf provides reason to speculate on the aetiology and pathogenesis of this lesion in the first place. It is possible that both the first, as well as the reformed ring or shelf, were acquired in postnatal life and

not present at birth (congenital). Such a view is supported by the fact that no experienced cardiac pathologist has reported this lesion causing critical obstruction in the newborn or infant human heart.

Experimentally, in a special strain of Newfoundland dogs a fully developed fixed subaortic ring/crescent was only found in dogs over 3 months.¹⁸ The puppies who died between the ages of 3 and 6 months showed the effects of endocarditis on the jet lesion of the aortic valve and not a severe obstructing lesion with secondary myocardial changes. In the only heart examined from a child under 2 months who was alleged to have died from fixed subaortic stenosis, personal inspection confirmed the presence of a fleshy shallow ridge just below the abnormal aortic valve, which did not cause serious obstruction with only a 1 to 2 mm protrusion. However, the heart had extreme thickening of the left ventricle and visible severe septal bulging, suggesting that the myocardium was abnormal and disproportionately abnormal in comparison to the mild fixed obstruction.

If it is accepted that the fixed subaortic stenosis removed by surgeons is acquired, there then occurs the problem of why it forms at all. That it forms in relation to some congenital abnormality of the outflow tract is likely, since 60 per cent of patients have other obvious congenital cardiovascular anomalies.

The pathological behaviour of the myocardium, including that with a "congestive" response, after operation for fixed subaortic stenosis in about 60 per cent raises the possibility that the primary abnormality is in the heart muscle and that this contracts abnormally causing turbulence in the outflow, with deposition of fibroelastic material which ultimately causes a fixed obstruction. Not only is there a clinical similarity in the behaviour of the left ventricle in the living patients with hypertrophic cardiomyopathy and some with fixed subaortic stenosis but also histological overlap as well as differences.^{19 20} Though the myocardium in these patients may be abnormal at birth it is doubtful if it is the cause of fixed subaortic stenosis as there are 30 to 40 per cent of patients who show no such problems after resection of fixed subaortic stenosis. Histological study of the muscle biopsied from the ventricular septum has been of no help in predicting the outcome since the biopsies are usually reported to show non-specific fibrosis and hypertrophy, even when there is good evidence of dynamic obstruction and unusual muscle. Perhaps this is because the biopsies are too superficial and the real problem lies deep in the septum and elsewhere. Helpful evidence comes from examination of two hearts, provided by Dr Luis Becú, from

children aged 9 and 15 months who died from lung infections, associated with a large duct. Trivial fixed subaortic stenoses were present in both; dysplasia, fibrosis, and hypertrophy were present in both but in neither was there enough dysplasia to consider that a primary myocardial abnormality was present.

One is therefore forced to search for some other primary (congenital) defect. In both hearts there was a minute non-obstructive fleshy ridge beneath the aortic valve cusp and the anterior mitral cusp at the site where the mature fixed subaortic stenoses are later found in some (Fig. 16). This is not the lesion found later and it resembled the fibrocartilagenous lump in the Newfoundland puppies.^{18 21} Perhaps this ridge is the remnant of some embryonic fold and is the basic congenital abnormality causing turbulence and the development of the mature fixed fibroelastic subaortic stenosis which the surgeon removes later. Such a course would be encouraged by the excess turbulence caused by the increased flow by a duct or ventricular septal defect present in many patients

with fixed subaortic stenosis. This fleshy ridge is not seen at operation nor in necropsy specimens from teenagers and adults who die with this disease, and is perhaps absorbed or sculptured out in natural changes in postnatal cardiac morphology. It could be argued that these hearts might never have developed fixed subaortic stenosis had the patients survived. We think it is likely they would and suspect that the little lump is the one congenital defect responsible for the development of the fully formed fixed subaortic stenosis which the surgeon removes many years later. If obstruction within the left ventricular outflow tract remains, turbulence persists and the same fibroelastic shelf or ring may form. The recent intriguing work of Rosenquist and his colleagues⁸ shows muscle bands causing increased separation between the aortic and mitral cusps in patients with fixed subaortic stenosis which may provide the missing link about the primary congenital abnormality.

Whatever the pathogenesis of subaortic stenosis or the cause of the abnormal muscle response, current policy is to remove fixed subaortic stenosis as soon as it is recognised, even with low gradients, that is below 30 mmHg, and not wait for secondary changes, or for symptoms, or for any electrocardiographic changes. The need to differentiate such patients from mild aortic valve stenosis is obvious since those with mild to moderate aortic valve stenosis should not have premature surgery as it may interrupt growth of the root or cause serious incompetence of the valve. Because of the difference in management any patient suspected of having fixed subaortic stenosis should be investigated with angiography early. Early experience suggests that the two-dimensional echo is reliable in recognising developed fixed subaortic stenosis, whereas the M-mode is not always, particularly in mild lesions. Furthermore, persistence of a long ejection systolic murmur in patients after duct ligation demands early investigation.

From careful evaluation of this group of patients late after resection of subaortic stenosis we conclude that results may not be as good as initial assessment suggests. The left ventricle may continue to behave abnormally either hypercontractile like hypertrophic myopathy, or "congestive" features and changes may advance. Patients need careful supervision, good early routine postoperative haemodynamic evaluation, and protection from infective endocarditis for life. Their problems are not completely cured and some may persist or even return mysteriously.

We thank Mr Keith Ross for permission to include the 10 patients on whom he operated in the National Heart Hospital. We are particularly grateful to



Fig. 16 Cross-section of heart from 9-month-old child with duct who died from overwhelming infection. This shows a minute subaortic ridge (arrow), not causing any important obstruction, beneath a normal aortic valve cusp (C). The ascending aorta (A) is normal.

Dr Luis Becú for providing information on the histology of fixed subaortic stenosis and two hearts for examination. We are indebted to Dr A Rickards who provided routine haemodynamic data and encouraged the extensive evaluation of left ventricular function, and to Dr Simon Rees and the late Dr Keith Jefferson for radiological evaluation.

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